Screening for childhood strabismus by primary care physicians

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ABSTRACT

OBJECTIVE To review the clinical classification of strabismus, to describe the timing and method of strabismus screening examinations, and to discuss the principles of treatment.

QUALITY OF EVIDENCE Current literature (1983 to 1995) was searched via MEDLINE using the MeSH headings strabismus, ocular motility disorders, and amblyopia. Articles were selected based on their date of publication, clinical relevance, and availability. Preference was given to more recent articles, articles with large numbers of subjects, and well-designed cohort studies. Official recommendations from academic groups were analyzed. Descriptions of clinical tests and their illustrations are based on classic texts.

MAIN FINDINGS Primary care physicians should screen all low-risk children. High-risk children (low birth weight, family history of strabismus, congenital ocular abnormality, or systemic conditions with vision-threatening ocular manifestations) should be referred to an ophthalmologist for screening. Screening should be performed in the neonatal period, at 6 months, and at 3 years (Grade A recommendation), as well as at 5 to 6 years (Grade B recommendation). Screening examination includes inspection, examining visual acuity, determining pupillary reactions, checking ocular alignment, testing eye movements, and ophthalmoscopy.

CONCLUSIONS Primary care physicians are essential to early detection of strabismus and amblyopia. Early detection can help minimize visual dysfunction, allow for normal development of binocular vision and depth perception, and prevent psychosocial dysfunction.

RÉSUMÉ

OBJECTIF Passer en revue la classification clinique du strabisme, décrire le calendrier et la méthode utilisée pour l'examen de dépistage du strabisme et discuter des principes du traitement.

QUALITÉ DES PREUVES Recension de la littérature (1983 à 1995) dans MEDLINE en utilisant les mots clés «strabismus», «ocular motility disorders» et «amblyopia». Les critères de sélection des articles furent : la date de publication, la pertinence clinique et la disponibilité. La préférence a été accordée aux articles plus récents, aux études comportant un nombre élevé de patients et aux études de cohortes démontrant une excellente méthodologie de recherche. On a également analysé les recommandations officielles de groupes d'universitaires. Les descriptions des tests cliniques et les illustrations sont tirées des volumes de référence classiques.

PRINCIPAUX RÉSULTATS Les médecins de première ligne devraient soumettre au dépistage tous les enfants à faible risque. Quant aux enfants à haut risque (petit poids à la naissance, antécédents familiaux de strabisme, anomalie oculaire congénitale, affections systémiques comportant des manifestations oculaires à risque pour la vision), le dépistage devrait être confié à un ophtalmologiste. Le calendrier des examens de dépistage est le suivant : période néonatale, six mois et trois ans (recommandation A) ainsi qu'à cinq ou six ans (recommandation B). L'examen de dépistage comprend l'inspection, l'examen de l'acuité visuelle et des réactions de la pupille, la vérification de l'alignement et des mouvements oculaires, ainsi que l'ophtalmoscopie.

CONCLUSIONS Il est essentiel que les médecins de première ligne participent à la détection précoce du strabisme et de l'amblyopie. La détection précoce peut contribuer à minimiser la dysfonction visuelle, à faciliter le développement normal de la vision binoculaire et de la vision stéréoscopique et à prévenir les dysfonctions psychosociales.

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CME

Screening for childhood strabismus by primary care physicians

Strabismus, or abnormal ocular alignment, is one of the most common eye problems encountered in children. It affects approximately 4% of children younger than 6 years[1] and is an important cause of visual and psychological disability.

Untreated, 30% to 50% will develop amblyopia, or lose vision permanently, in the misaligned eye.[2] Treatment of amblyopia is rarely successful in restoring vision if instituted after age 8 to 10 years.[3] Furthermore, ocular alignment is necessary for normal development of binocular vision and depth perception.

Strabismus has many causes. Although most cases in childhood are primary or idiopathic, strabismus can indicate serious ocular disorders (eg, cataract, retinoblastoma). For these reasons, early detection and treatment of strabismus are essential.

When strabismus is present, each eye fixates on different objects at different points in space. The two dissimilar images cannot be fused and binocular vision is impossible. In children, the brain quickly learns to suppress the unfocused image from the deviated eye. This can result secondarily in clear vision in the one straight eye and decreased vision (amblyopia) in the deviated eye. The clear monocular image is inadequate for normal development of stereoscopic (three-dimensional) depth perception.

Ocular misalignment can be manifest (heterotropia) or latent (heterophoria). In heterophoria, the ocular deviation is kept latent by the fusional mechanism of the brain. That is, the eyes are aligned when both eyes are open. With fatigue, illness, or stress, the deviation becomes apparent. As well, covering one eye will interrupt fusion and result in a deviation of the eye under the cover. This tendency is the basis of the cover-uncover test. Most small latent deviations are asymptomatic. However, large latent deviations can result in asthenopia (eye strain), headache, or transient diplopia (double vision).

In heterotropia, the ocular misalignment is manifest because the fusional mechanism is unable to control the deviation. The position of the deviated eye is specified by the prefixes eso- (inward, nasal, convergent), exo- (outward, temporal, divergent), hyper- (upward, superior), and hypo- (downward, inferior). The type of strabismus is specified by the suffixes -phoria (latent) and -tropia (manifest). Esotropia is commonly referred to as "crossed eyes" and exotropia as "wall-eyes."

The peak age of onset of strabismus is birth to 4 years.[4] The most common form of childhood strabismus is esotropia, which accounts for well over 50% of all ocular deviations.[5] Exotropia is the second most common form and accounts for 25%. Most cases of childhood strabismus are concomitant. In concomitant strabismus, the degree of deviation is equal in all directions of gaze. In contrast, most adult-onset cases of strabismus are paralytic, in which the degree of deviation varies with the direction of gaze. Paralytic strabismus is usually caused by extraocular muscle paralysis (such as third nerve palsy in diabetes) or restriction (such as Grave's disease or orbital fracture with muscle entrapment). This paper focuses on concomitant strabismus in children.

This article provides a clinical classification of strabismus, describes recommended ocular screening examinations, and discusses principles of treatment. The aim is to refresh the knowledge of primary care physicians about early detection of strabismus. The ultimate goal is to reduce the incidence of preventable vision impairment resulting from amblyopia.

Literature search

A MEDLINE search from 1983 to 1995 was conducted using the MeSH headings strabismus, ocular motility disorders, and amblyopia. The search was limited to English-language articles. Articles were selected based on their date of publication, clinical relevance, and availability. Preference was given to more recent articles, studies with large numbers of subjects, and well-designed cohort studies. In addition, we used bibliographies of the articles to find additional articles. Several textbooks on eye care for primary care physicians,[1] on general ophthalmology,[5,6] and on strabismus were used.[7]

The recommendations of the Canadian Task Force on the Periodic Health Examination, the US Preventive Services Task Force, the American Academy of Pediatrics, and the American Academy of Ophthalmology were analyzed. Most of the recommendations in this article are based on the recommendations of these official bodies. The description of the various clinical tests and their illustrations are based on classic texts.

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Causes

Most cases of childhood concomitant strabismus are primary or idiopathic. In primary strabismus, the extraocular muscles are normal in structure and function, and no ocular organic disorder can be found.[7] There is often a family history of strabismus. Also, strabismus can be secondary to errors of refraction, such as anisometropia and marked hyperopia, or other ocular organic disorders.

In anisometropia the refractive power of the two eyes is quite different. The brain receives a blurred image from one eye and a clear image from the other. The eye with the clear image remains aligned on target; the blurred eye can deviate. The direction of deviation is most often esotropic in children with anisometropia.

Hyperopic or far-sighted individuals must accommodate (focus or increase their power of refraction) for both distant and near vision in order to see clearly. Accommodation and convergence normally occur together. Individuals with marked hyperopia must make a large effort to see, which sometimes leads to excessive convergence resulting in esotropia, known as accommodative esotropia.[8] Eye misalignments can also be caused by excessive divergence drive leading to exotropia or vertical imbalances leading to hypertropias.

Strabismus can also be caused by ocular disease. Conditions that compromise vision in one or both eyes can result in ocular misalignment. Such conditions include corneal opacities, cataracts, retinoblastoma, and tumours of the optic nerve. Although these conditions are rare, it is essential to detect them early.

Assessment

The timing and frequency of screening examinations for strabismus are not well established. The Canadian Task Force on the Periodic Health Examination suggests repeat examination of the eyes for strabismus at well-baby visits (in the neonatal period and at 6 months)[9] with a Grade A (good evidence for) recommendation. The Task Force suggests eye and vision testing in the preschool period (3 years and 5 to 6 years)[9] with a Grade B (fair evidence for) recommendation. The American Academy of Ophthalmology[10] and the American Academy of Pediatrics[11] make similar recommendations.

The limitations of screening tests have been studied. Specially trained nurses screened 59 782 preschoolers. Using visual inspection and assessment of visual acuity and of ocular alignment, the screening tests had a negative predictive value of 98.7% for amblyopia, strabismus, and high refractive error.[12] A similar program had a positive predictive value of 72%.[13] Despite suboptimal performance of the screening tests, early detection and treatment of strabismus is essential to prevent important visual and psychosocial dysfunction.

Table 1. Screening tests for eye disease in children

AGE	EYE SCREENING BY PRIMARY CARE PHYSICIAN
Neonate	External examination Ocular alignment Corneal light reflex test Ophthalmoscopy Clear media Normal red reflex
6 months	Above tests plus: Ocular motility Fix upon and follow a small toy with each eye Pupillary reactions
3 years	Above tests plus: Visual acuity Sheridan Gardner (single-letter matching game) or picture chart Cover test
5-6 years	Above tests plus Snellen visual acuity

The essential components of strabismus screening have not been established by clinical trials. The American Academy of Pediatrics recommends[11] the following (**Table 1**). Neonatal assessment should include external examination of the eye and surrounding tissues, corneal light reflex test for ocular alignment, and examination for red reflex. Examination at 6 months should include all of these tests and assessment of ocular motility (ability to fixate upon and follow a small toy with each eye) and pupillary reactions. At 3 years, one assesses visual acuity (using a picture chart or a Sheridan Gardiner chart), ocular alignment (by corneal light reflex test and cover test), and ocular motility (tested by gaze in the six cardinal fields). External examination, pupillary reactions, and ophthalmoscopy are also performed. Screening at age 5 to 6 years is similar to the 3-year examination, but visual acuity is assessed using a Snellen chart.

All children should be screened for strabismus. Primary care physicians should screen all low-risk

Tests of ocular alignment[14]

Inspection

Casual observation of a child's ocular alignment works reasonably well for large deviations. However, it is unreliable for small or moderate deviations. As well, many young children have the appearance of crossed eyes despite straight ocular alignment (pseudostrabismus).

Corneal light reflex (Hirschberg) test

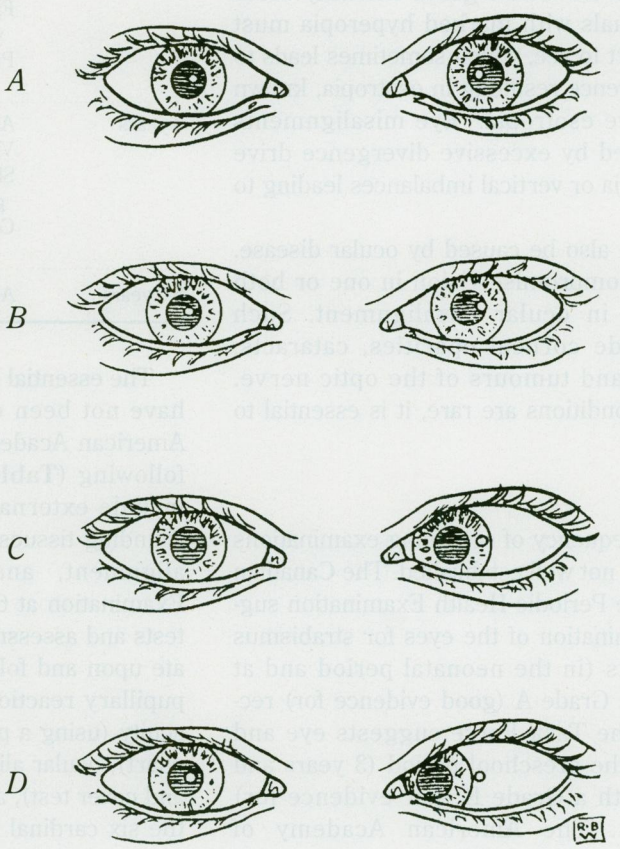
This test (**Figure 1**[15]) provides a rapid and easy method to detect manifest strabismus. The examiner projects a light source (eg, penlight) onto the cornea of both eyes simultaneously from a distance of approximately 70 cm. The child is encouraged to look at the light. The examiner then compares the position of the corneal light reflex in each eye. In straight eyes, the light reflex appears symmetric and central (or very near central) in both eyes. If strabismus is present, the reflected light is asymmetric and appears off-centre in one eye. In esotropia, the light reflex appears temporally deviated. In exotropia, the light reflex appears nasally deviated.

Cover tests

Cover tests require patient cooperation, good eye movement capacity, and reasonable visual acuity. Thus, it is extremely difficult to perform a cover test on children younger than 1 year. These tests are the most accurate method of evaluating ocular alignment and can be performed for both distant and near fixation.

Cover test: The child fixates on a target with both eyes (**Figure 2**[15]). The examiner places an occluder over one eye. The unoccluded eye is observed for movement. An inward movement of the unoccluded eye indicates exotropia (divergent strabismus) in that eye. When the

Figure 1. Hirschberg light reflex test: *A) corneal light reflexes are in a normal position of slight nasal decentration. B-D) Light reflex is displaced progressively more temporally in the left eye, indicating increasing amounts of left esotropia.*



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good eye is covered, the exotropic eye moves to centre (inward) to assume fixation. An outward movement of the unoccluded eye indicates esotropia (convergent strabismus) in that eye. The cover test is then performed upon the second eye.

Cover-uncover test: The child fixates on a target with both eyes (**Figure 3**[15]). The examiner places an occluder over one eye (interrupting fusion). The occluder is then removed and the examiner observes for movement in the newly uncovered eye. Inward movement of the newly uncovered eye indicates exophoria in that eye. When the exophoric eye is covered, fusion is broken and the eye turns out. When that eye is uncovered, fusion is restored and the eye turns to centre to resume fixation. An outward movement of the newly uncovered eye indicates esophoria in that eye.

Figure 2. Cover test: *A) Position of patient's eyes before the test (in normal results). B) Cover placed over left eye does not elicit a fixation movement of the right eye (normal results: right eye does not deviate). C) Cover placed over right eye does not elicit a fixation movement of the left eye (normal results: left eye does not deviate). D) Right eye moves outward to fixate when left eye is covered (right esotropia). E) Right eye moves inward to fixate when left eye is covered (right exotropia).*

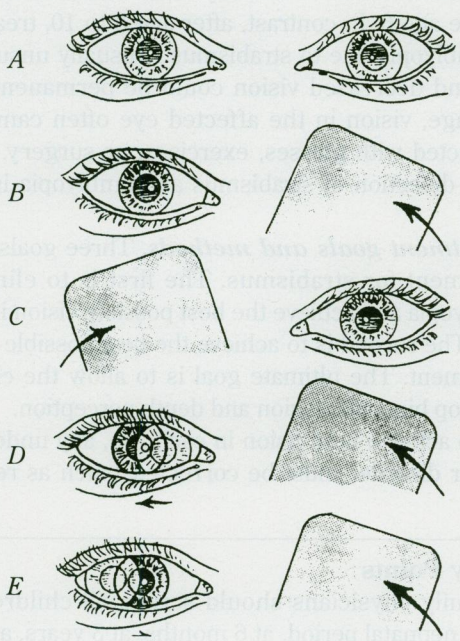

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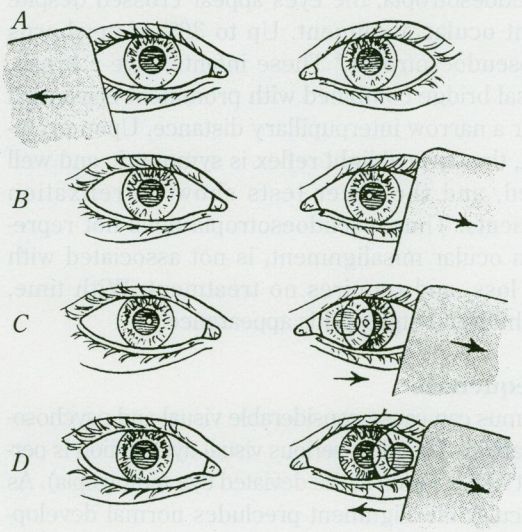
Figure 3. Cover-uncover test: *A) Cover has been removed from right eye, and no movement can be detected (normal results: no latent deviation of right eye). B) Cover has been removed from left eye, and no movement can be detected (normal results: no latent deviation of left eye). If conditions in A and B are present, the patient has no phorias that can be detected with this test. C) When uncovered, left eye moves outward to fixate (esophoria). D) When uncovered, left eye moves inward to fixate (exophoria).*



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children. High-risk children should be referred to an ophthalmologist for screening. Factors that place children at high risk for developing strabismus are listed in **Table 2**.[1]

The American Academies' recommendations were based on "the best available scientific data as interpreted by panels of knowledgeable health professionals."[10] Because the studies were evaluated by panels of specialists, their guidelines could be overly inclusive.

Essential components of the ocular screening examination, as appropriate for age, are inspection, visual acuity, pupillary reactions, ocular motility, corneal light reflex test, cover tests, and ophthalmoscopy (**Table 1**).

Table 2. Risk factors for developing strabismus

Low birth weight (< 1250 g)
Family history of strabismus
History of congenital ocular abnormality (eg, infantile cataracts, retinoblastoma) or of systemic conditions with vision-threatening ocular manifestations (eg, pauciarticular juvenile rheumatoid arthritis, which can cause iritis and cataracts)

Pseudostrabismus

In pseudoesotropia, the eyes appear crossed despite straight ocular alignment. Up to 30% of newborns have pseudoesotropia.[4] These infants have a broad, flat nasal bridge combined with prominent epicanthal folds or a narrow interpupillary distance. Upon examination, the corneal light reflex is symmetric and well centred, and the cover tests show no refixation movements. Thus, pseudoesotropia does not represent an ocular misalignment, is not associated with vision loss, and requires no treatment. With time, most children outgrow this appearance.

Consequences

Strabismus can cause considerable visual and psychosocial disability. The most serious visual dysfunction is permanent loss of vision in the deviated eye (amblyopia). As well, ocular misalignment precludes normal development of binocular vision and depth perception.[16] Children with strabismus sometimes have difficulty participating in ballgames with friends and classmates.[17] This can lead to isolation at school, poor self-esteem, and social dysfunction. Children often endure the teasing of classmates and the misguided frustration of teachers ("Why don't you look at me when you are speaking?"). Furthermore, abnormal binocular vision can interfere with visual motion processing[18] and with reading.[19]

Adults with uncorrected or incompletely corrected childhood strabismus report difficulties with self-image, securing employment, relationships, school, work, and sports.[20] Affected individuals sometimes have difficulty qualifying for certain classes of driver's licences and for pilot's licences.[17] Thus, strabismus can cause psychosocial problems in addition to dysfunctional vision.

Approach to management

When to refer. Primary care physicians should screen all low-risk children for strabismus. Those with risk factors (**Table 2**) should be referred to an ophthalmologist for screening. If a manifest ocular misalignment is detected, the child should be referred to an ophthalmologist for further management. A latent ocular misalignment requires no treatment if asymptomatic but should be closely followed by a primary care physician. If symptoms such as eyestrain, double vision, or headaches develop, children should be referred to an ophthalmologist for further assessment and management.[10] If an ophthalmologist is not accessible, evaluation by an optometrist experienced with children's eye problems is acceptable.

Treatment of amblyopia due to strabismus or refractive error is usually successful if initiated before age 6. In contrast, after age 8 to 10, treatment of amblyopia due to strabismus is usually unsuccessful, and decreased vision could be permanent.[21] By this age, vision in the affected eye often cannot be corrected with glasses, exercises, or surgery. Thus, early detection of strabismus and amblyopia is vital.

Treatment goals and methods. Three goals guide treatment for strabismus. The first is to eliminate amblyopia and achieve the best possible vision in each eye. The second is to achieve the best possible ocular alignment. The ultimate goal is to allow the child to develop binocular vision and depth perception.

To achieve best vision in each eye, any underlying ocular disorder must be corrected (such as removal

Key Points

Family physicians should screen all children in the neonatal period, at 6 months, at 3 years, and at 5 or 6 years. Screening includes inspection, examining visual acuity, determining pupillary reactions, checking ocular alignment, testing eye movements, and ophthalmoscopy.

of congenital cataracts), and any serious refractive error must be corrected (such as that associated with esotropia). Accompanying amblyopia must be vigorously treated using occlusive therapy (patching the "good" eye to force the child to use the "lazy" eye).[3]

Ocular alignment must be restored for normal binocular vision and depth perception to develop. Non-surgical treatment options include glasses, eye exercises, and prisms. Strabismus surgery or botulinum toxin injection are options if significant residual deviation remains after use of non-surgical therapies or if such therapies are not indicated. Ocular realigning procedures for congenital strabismus are best performed before the age of 2 years to maximize chances for development of binocular vision.[22] Surgery for other forms of childhood strabismus is generally recommended before the child is 6 to 8 years old. A secondary goal of ocular alignment procedures is to improve cosmesis, in order to minimize psychosocial distress associated with strabismus.

Conclusion

Strabismus is a common problem affecting 4% of school-age children. It has important visual and psychosocial sequelae. Untreated, up to 50% of patients with heterotropias will develop permanent vision loss in the deviated eye. Improperly aligned eyes will not allow normal binocular vision or stereoscopic depth perception to develop. This can interfere with a child's ability to read, to play sports, and to relate to others. This social dysfunction persists into adulthood in its effect on self-image, employment, and relationships.

Primary care physicians should screen all children for strabismus in the neonatal period, at 6 months, at 3 years, and at 5 to 6 years. Essential screening components include inspection, assessing visual acuity, determining pupillary reactions, testing ocular motility, corneal light reflex test, cover tests, and ophthalmoscopy. All cases of manifest strabismus and all symptomatic cases of latent strabismus should be referred to an ophthalmologist promptly. Amblyopia can be successfully treated, and binocular vision and depth perception can develop normally if strabismus and amblyopia are detected and treated early. In cases of serious ocular disease, such as retinoblastoma, lives can be saved. Primary care physicians play an essential role in early detection of these conditions.

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References

1. Trobe J. *The physician's guide to eye care.* San Francisco: American Academy of Ophthalmology; 1993. p. 18-19, 157.
2. Lavrich J, Nelson L. Diagnosis and treatment of strabismus disorders. *Pediatr Clin North Am* 1993;40(4):737-52.
3. Watts J. Squint. *Practitioner* 1989;223(1479):1553-6.
4. Catalano J. Strabismus. *Pediatr Ann* 1990;19(5):289,292-7.
5. Asbury T, Burke MJ. Strabismus. In: Vaughan D, Asbury T, Riordan-Eva P, editors. *General ophthalmology.* East Norwalk, Conn: Appleton & Lange; 1992. p. 231-50.
6. Nelson L, Catalano R. *Atlas of ocular motility.* Philadelphia: WB Saunders; 1989.
7. Parks M, Wheeler M. Concomitant esodeviations. In: Tasman W, Jaeger G, editors. *Duane's clinical ophthalmology.* Philadelphia: JB Lippincott; 1991. p. 1-14.
8. Caloros E, Rouse M. *Clinical management of strabismus.* Boston: Butterworth-Heinemann; 1993.
9. Canadian Task Force on the Periodic Health Examination. *Canadian guide of clinical preventative health care.* Ottawa: Ministry of Supply & Services; 1994. p. 258 66, 298-304.
10. The American Academy of Ophthalmology. *Policy statement—vision screening for infants and children.* San Francisco: The American Academy of Ophthalmology; 1996.
11. The American Academy of Pediatrics, Committee on Practice and Ambulatory Medicine. Vision screening and eye examination in children. *Pediatrics* 1986;77:918-9.
12. De Becker I, MacPherson H, LaRoche G, Braunstein J, Cottle R, McIntyre L, et al. Negative predictive value of a population-based preschool vision screening program. *Ophthalmology* 1992;99(6):998-1003.
13. MacPherson H, Braunstein J, LaRoche G. Utilizing basic screening principles in the design and evaluation of vision screening programs. *Am Orthopt J* 1991;41:110-21.
14. Deutsch J, Nelson L. Diagnosis and management of childhood strabismus. *Pediatrician* 1990;17(3):152-62.
15. Von Noorden GK. *Atlas of strabismus.* 4th ed. Toronto: C.V. Mosby Company; 1983.
16. Manny R, Martinez A, Fern K. Testing stereopsis in the preschool child. *J Pediatr Ophthalmol Strabismus* 1991;28:223-31.
17. Pratt-Johnson J, Tillson G. *Management of strabismus and amblyopia.* New York: Thieme Medical Publishers; 1994. p. 11-4.
18. Tychsen L, Lisberger S. Maldevelopment of visual motion processing in humans who had strabismus with onset in infancy. *J Neurosci* 1986;6(9):2495-508.
19. Simons H, Grisham J. Binocular anomalies and reading problems. *J Am Optom Assoc* 1987;58(7):578-87.
20. Satterfield D, Keltner J, Morrison T. Psychosocial aspects of strabismus study. *Arch Ophthalmol* 1993;111(8):1100-5.
21. Rubin S, Nelson L. Amblyopia—diagnosis and management. *Pediatr Clin North Am* 1993;40(4):727-35.
22. Ing M. Early surgical alignment for congenital esotropia. *J Pediatr Ophthalmol Strabismus* 1983;20:11-8.